Treatment of Brooke-Spiegler Syndrome Trichoepitheliomas with Erbium:Yttrium-Aluminum-Garnet Laser: A Case Report and Review of the Literature

ABSTRACT

Patients with Brooke-Spiegler Syndrome (BSS) can present with benign cylindromas, spiradenomas, spiradenocylindromas, and trichoepithelioma. Therapy options include excision, electrocautery, CO_2 laser ablation, dermabrasion, and radiofrequency. Here, we present a patient with BSS with multiple trichoepitheliomas who was successfully treated with erbium:yttrium-aluminum-garnet (YAG) laser therapy and review similar cases of BSS treated with a YAG laser modality.

KEY WORDS: Brooke-Spiegler syndrome, trichoepithelioma, erbium:YAG laser

by LOGAN W. THOMAS, MD; CHRISTINE T. PHAM, BS; BRANDON COAKLEY, MD; and PATRICK LEE, MD

All authors are with the Department of Dermatology, University of California, Irvine, in Irvine, California.

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CASE REPORT

A 28-year-old female patient presented to the clinic with multiple skin-colored papules on her face that had been present for 15 years prior to visiting the clinic. The patient reported that her mother, maternal uncles, and maternal cousins had similar lesions on their face, scalp, and body.

Dermatologic examination showed multiple agminated flesh-colored 2- to 5-mm dome-shaped papules predominantly within the alar grooves, base of the alar rim, and within the nasolabial folds (Figure 1). Two reddish-colored nodules were also identified on the vertex and lateral aspect of the scalp. A deep-seated dermal nodule was additionally noted on the right breast. Three skin biopsies were performed and histopathological examination suggested a cylindroma on the scalp, a spiradenoma on the breast, and a trichoepithelioma on the lip. The combination of diagnostic criteria was necessary to confirm the diagnosis, with genetic testing suggested, though this was not performed in our patient (Table 1). Based on the clinical history, family history, and histopathologic findings, a diagnosis of Brooke-Spiegler syndrome (BSS) was made.

The patient was recommended to undergo Er:YAG laser therapy using the Sciton profractional laser (Sciton, Palo Alto, California). The treatment area was anesthetized by the combination of a topical anesthetic and nerve blocking with 1% lidocaine and epinephrine. The Er:YAG laser was set at the setting of 50µ and an ablation depth at 10 Hz, with suction and no coagulation. The tumors were ablated to the depth of the papillary dermis and no complications were noted aside from expected erythema and tenderness. At both the one- and 31-month follow-up, no recurrence was observed (Figures 2, 3, 4).

DISCUSSION

BSS is a rare autosomal dominant genetic

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CORRESPONDENCE: Christine T. Pham, BS; Email: christinethpham@gmail.com

FIGURE 1. Right and left sides of the face of a patient with multiple trichoepitheliomas on the bilateral nasolabial fold, upper lip, and alar rim

FIGURE 2. Right and left sides of the face after the removal of the multiple trichoepitheliomas with mild erythema at one month after treatment with Er:YAG laser

disorder marked by the development of cutaneous neoplasms, such as cylindroma, spiradenoma, spiradenocylindroma, and trichoepithelioma. The syndrome results from a gene mutation in the cylindromatosis gene (CYLD), located on chromosome 16q12-q13.[1] The cutaneous neoplasms are usually benign tumors but, because of the progressive nature of the condition, some reports detail findings of nodules becoming malignant, growing, and developing ulcers and bleeding.[2] Volter et al[3] observed that malignant transformation is more likely with multiple cylindromas and close follow-up is recommended.[3] The condition usually presents itself in young adults and worsens with age. The tumors are normally situated on the scalp, face, and neck area, measuring about 0.5 to 3cm in diameter and varying from a few to several hundred nodules.[1]

Different treatment modalities have been used to treat BSS, including excision, electrocautery, CO_2 laser ablation, dermabrasion, and radiofrequency.[4–7] Recurrence of the nodules are common and excessive scarring can occur. In this case, an Er:YAG laser treatment (10 Hz) was adopted with the following settings: 4mm spot size, 5 Joules, and 10 Hz. These settings allowed the laser surgeon to precisely and uniformly remove 250µ of tissue per second, which enabled gradual thinning of the target area. The entire procedure took approximately 10 minutes, during which time, the tumors were ablated in a meticulous and controlled manner. The Er:YAG laser was chosen due to research showing successful results with ablation and

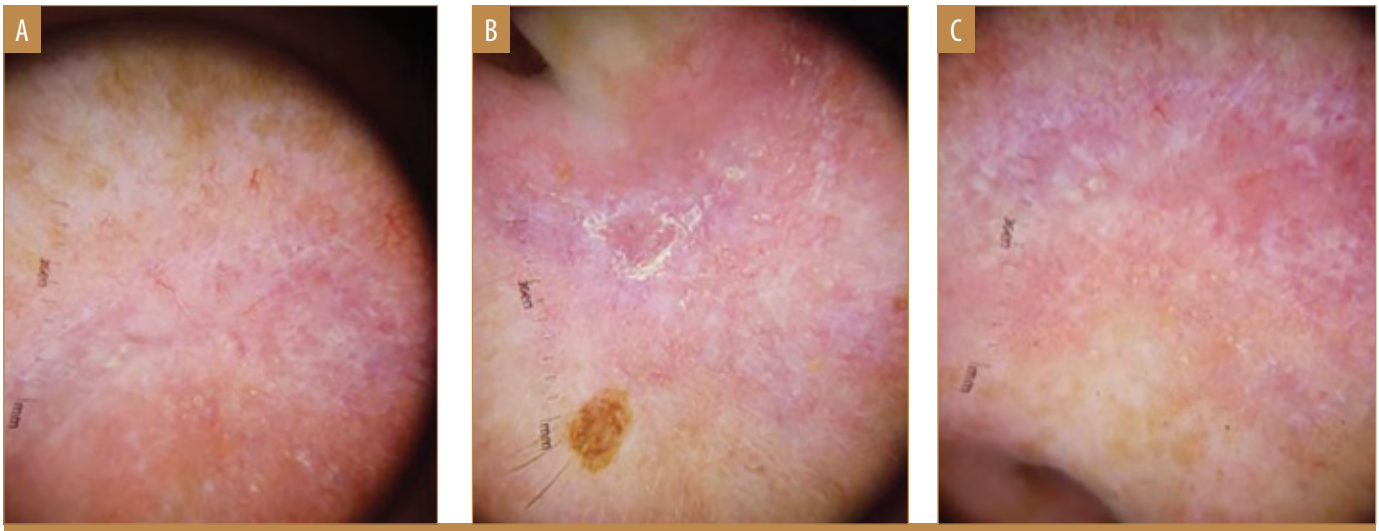

FIGURE 3. A: dermatoscopic image of the right side of the nose one month posttreatment with Er:YAG laser and; **B,C:** dermatoscopic images of the left nose one month posttreatment with Er:YAG laser

TABLE 1. Diagnostic requirements for Brooke-Spiegler Syndrome

DIAGNOSTIC CRITERIA
Early stage of onset for skin neoplasms
Family history of neoplasms with autosomal dominance inheritance pattern
Histopathologic findings on biopsy of cylindromas, spiradenomas, or trichoepitheliomas
Increased association with tumors in parotid or salivary glands
Genetic testing confirming mutation in CYLD gene

FIGURE 4. Right and left sides of the face demonstrating no recurrence of the multiple trichoepitheliomas 31 months after treatment with Er:YAG laser

contour and blend with surrounding tissue.[8,9] Also, using an Er:YAG laser facilitates the visualization of hamartomatous tissue while ablating due to a lack of the tissue carbonization normally seen with CO_2 lasers. This improved visualization prevents excessive damage and scarring to normal tissue.[5,7] The CO_2 laser beam is scattered within the cutaneous tissue layer, causing thermal coagulation necrosis not only in the deep tissues directly underneath the laser but also in the surrounding tissues. In contrast, the Er:YAG laser is more precise and does not affect the surrounding tissues like the CO_2 laser does. Thermal damage is minimized during the ablation process, leading to the reduction of scarring.

A review of the literature returned three case reports that have used either the Er:YAG laser or neodymium-doped:YAG laser for treatment of either cylindromas and trichoepitheliomas found in BSS (Table 2). One report detailed the successful treatment of trichoepitheliomas on the lip and nasolabial folds with use of an Er:YAG laser (2-mm beam diameter, 2940-nm wavelength, 0.4 J/cm^2 at 4 pulses/sec) for ablation, followed by CO_2 laser (2mm, 5 W) application for homeostasis, with no recurrence within two years.[5] Another case study of one patient compared the efficacy of Er:YAG laser resurfacing, photodynamic therapy (PDT) with 5-aminolevulinic acid, and imiquimod 5% cream in the treatment of multiple trichoblastoma of the face and found that Er:YAG combined with PDT therapy was slightly better than laser therapy alone. Ultimately, however, the patient favored laser therapy alone due to the ease of treatment despite combined PDT and laser therapy showing slightly better results.[7] Another report presented the successful treatment of a patient with dermal cylindroma of the scalp with a neodymium-doped:YAG laser. Bleeding was decreased in comparison with when using a CO_2 laser due to the neodymium-doped:YAG laser's ability to seal large blood vessels up to

TABLE 2. Review of available literature discussing the treatment of BSS with Er:YAG or neodymium-doped:YAG laser

STUDY DESIGN/ SAMPLE SIZE	CONDITION TREATED	INTERVENTION	NUMBER OF TREATMENTS	OUTCOME MEASURES	CLINICAL OUTCOMES	ADVERSE EFFECTS
Rallan et al.[5]						
Case report (n=1)	Trichoepithelioma in BSS	Continuum Biomedical™ Er:Yag (2-mm beam diameter, 2,940nm wavelength, 0.4 J/cm^2 at 4 pulses/sec) followed by Nidek™ CO_2 laser was used in continuous mode (2 mm, 5 W)	Every 3–4 months over 19 months	Recurrence at follow-up	No recurrence during two years of follow-up	Not reported
Lopiccolo et al.[7]						
Case report (n=1)	Trichoepithelioma in BSS	Each side of face was divided into three portions: the top one-third received PDT with 5-aminolevulinic acid, the middle one-third received topical imiquimod 5% cream, and the lower one-third was control; all three zones also received Er:YAG Sciton profractional laser at a depth of 450µm and a coagulation 3 setting, with 11% overlap therapy on the right side of the face	Two days after laser resurfacing, 1g of 5-aminolevulinic acid was evenly applied; after a one-hour incubation period, the area was treated with a Sciton broadband device with a 420-nm cutoff filter at 22 J/cm^2 with a 20-ms pulse width. Eight hours after PDT, a thin film of imiquimod 5% cream was applied. Patient was treated for three months, with laser resurfacing occurring once each month, PDT once every two weeks, and imiquimod cream applied every other night	Patient evaluation of cosmetic appearance pretreatment and three months posttreatment Rated on a scale of 0–10 points, where 0 points=no disease and 10 points=severe disease	Before treatment: 10 points; 3 months after treatment: 7 in the zone treated using PDT alone. 6 in Er:YAG resurfacing alone and resurfacing plus imiquimod. 5 in PDT and laser resurfacing. Patient preferred laser alone compared to PDT and laser	Not reported
Tarstedt et al.[8]						
Case report (n=1)	Cylindroma in BSS	Nd:YAG laser (1,064 nm, Sharplan 2,100, Gastrofiber 600 mm) used in a cutting continuous mode of 40 W	One treatment with excision with healing by secondary intention	Recurrence at follow-up	No recurrence during four years of follow-up	Not reported

BSS: Brooke-Spiegler syndrome; Er:YAG: erbium-doped yttrium-aluminum-garnet; Nd:YAG: neodymium-doped yttrium-aluminum-garnet; PDT: photodynamic therapy

2mm in diameter by internal heating of the cylindroma. In addition, no relapse occurred after more than four years.[10]

The relapse of nodules in BSS is a common occurrence. No relapse occurred during the patient's one- and 31-month follow-up appointments in this case and the results were deemed to be aesthetically pleasing. However, continuing to monitor patients for a longer period of time is necessary to deduce the effectiveness of Er:YAG laser application in preventing relapse. Furthermore, other treatment modalities should be explored. A case study involving CO_2 laser treatment and topical 1% sirolimus cream as suppressive therapy to treat patients with multiple trichoepitheliomas showed limited progression of the disease.[7] Another study compared neodymium-doped:YAG, Er:YAG, and CO_2 laser treatment and saw significant improvements in skin rejuvenation using the neodymium-doped:YAG laser.[11]

CONCLUSION

BSS is a rare condition that requires further investigation into proper lesion removal modalities and best results for the prevention of lesion recurrence. This case reports shows the effectiveness of Er:YAG laser therapy alone in the removal of facial trichoepitheliomas.

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